

Kaposi's Sarcoma in a Patient With Rheumatoid Arthritis and Polymyositis Treated With Corticosteroids

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KAPOSI'S SARCOMA is more prevalent in immunocompromised patients. It has been documented in patients with lymphoid malignant disorders,¹ renal allografts,^{2,3} and more recently in patients with the acquired immunodeficiency syndrome (AIDS).⁴ Evidence is mounting that Kaposi's sarcoma occurs with greater frequency in patients with autoimmune diseases. With the exception of the AIDS-associated cases, a common thread among the other high-risk groups is the concomitant use of corticosteroids with or without other immunosuppressive agents. We herein present the case of a man in whom Kaposi's sarcoma occurred in conjunction with rheumatoid arthritis and polymyositis, and we review the literature available on the association of Kaposi's sarcoma with autoimmune disorders and corticosteroid therapy.

Report of a Case

A 57-year-old man was admitted to UCLA Medical Center in 1965 for the diagnosis and treatment of Kaposi's sarcoma. Four years before admission the patient was diagnosed to have seropositive, nodular, erosive rheumatoid arthritis and Raynaud's phenomenon. He was given salicylates and low doses of prednisone. Three years later the patient had pronounced symmetric proximal muscle weakness, an elevated serum creatine kinase level to 2,003 units per liter (normal range 21 to 215), an abnormal electromyogram, and a muscle biopsy confirming a diagnosis of polymyositis. The biopsy specimen showed no evidence of steroid-induced myopathy. A good clinical response was obtained to treatment with 60 mg of prednisone a day. The creatine kinase level decreased to the normal range, and the proximal muscle strength increased. Two months before admission, multiple violaceous skin nodules developed on both lower extremities. The patient had no preexisting nodular skin lesions before treatment with high-dose corticosteroids.

On examination the vital signs and the head, eyes, ears, nose, and throat were normal. Lymphadenopathy was noted in the anterior cervical lymph node chain. Rales were heard in the posterior lower lobes of the lung. Examination of the heart revealed a murmur of aortic sclerosis confirmed by echocardiography, the liver was palpated 5 cm below the right costal margin, and the findings of the abdominal examination were otherwise normal. Dependent edema was noted in the lower extremities, along with multiple small—2 to 6

mm—round, discrete, violaceous nodules on the skin (Figure 1).

The musculoskeletal examination revealed symmetric synovitis in the proximal interphalangeal joints, metacarpal phalangeal joints, elbows, and knees. Both olecranon and prepatellar bursae were enlarged. A 0.5-cm nodule was discovered in proximity to the left elbow. Muscle testing showed 4/5 strength in all proximal muscles.

Laboratory studies elicited the following values: hemoglobin, 10.5 grams per dl; leukocyte count, 7,400 per μ l with a normal differential; erythrocyte sedimentation rate, 53 mm per hour (Wintrobe); and elevated levels of serum aspartate aminotransferase (glutamic-oxaloacetic transaminase), lactic dehydrogenase, and creatine kinase. A urinalysis disclosed trace amounts of protein, but was otherwise normal. The rheumatoid factor was 1:160, tests for the antinuclear antibody and lupus erythematosus preparation were negative, and the third and fourth component of complement and total hemolytic complement values were also normal. The diagnosis of Kaposi's sarcoma was confirmed by a skin biopsy, with the presence of rheumatoid arthritis confirmed both by biopsy of the nodule and typical clinical and radiologic findings. The diagnosis of polymyositis was reconfirmed by the clinical presentation, elevated muscle enzyme values, classic electromyographic findings, and deltoid muscle biopsy findings. Bibasilar interstitial infiltrates were present on chest radiographs. Liver enlargement was thought to be due to passive congestion and lymphadenopathy related to rheumatoid arthritis with local joint involvement.

Radiation treatment of the skin lesions was considered, but before treatment could be initiated, the patient was discharged to his primary physician and died six months later of heart failure complicated by pulmonary insufficiency.

Discussion

Kaposi's sarcoma was first termed "multiple idiopathic pigmented hemangioma" by Moritz Kaposi in 1872.⁵ The classic presentation of Kaposi's sarcoma is that of one or more skin lesions occurring predominantly on the extremities in elderly men of Jewish or Italian lineage.⁶ It is now recognized to have an increased prevalence in immunocompromised hosts. With regard to autoimmune diseases, Kaposi's sarcoma has been reported in patients with rheumatoid arthritis,⁷⁻⁹ systemic lupus erythematosus,^{10,11} dermatomyositis,¹² polymyositis,^{13,14} polymyalgia rheumatica,⁸ and temporal arteritis.¹⁵ All of these cases occurred in patients receiving corticosteroids systematically. In a recent case report, Kaposi's sarcoma developed in a homosexual man with Henoch-Schönlein purpura after six months on oral prednisone therapy.¹⁶ Our patient is unique for having two connective tissue disorders with the subsequent development of Kaposi's sarcoma during corticosteroid therapy. The presence of rheumatoid arthritis and polymyositis in this patient was well documented by clinical, serologic, radiographic, and histopathologic findings.

Review of these cases has not shown a clear time- or dose-dependent relationship between corticosteroid treatment and the onset of Kaposi's sarcoma lesions, although the duration of steroid therapy in most cases was at least four months. Spontaneous tumor regression has been reported with complete clinical remission in certain patients after discontinuing the corticosteroid therapy or other immunosuppressive agents.^{12,15,16}

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The mechanism by which Kaposi's sarcoma arises in patients receiving corticosteroids is not well understood. Postulated mechanisms have included the activation of latent oncoviruses as a result of chronic immunosuppression, in addition to the presence of aberrant immune responses related to the underlying disease.^{2,8,10} Genetic influences may also play a role in the development of this particular neoplasm in immunocompromised persons.

Our patient had Kaposi's skin lesions in a distribution more typically seen in the "classic" form of the disease—that is, confined to the skin and localized to the extremities. The variety of Kaposi's sarcoma seen in association with AIDS more commonly presents with lesions on the torso as well and can occur in disseminated forms with significant visceral involvement. Visceral involvement occurs in as many as 10% of cases of the classic variety, and, to date, only one case of disseminated Kaposi's has been associated with corticosteroid therapy in a patient with systemic lupus erythematosus.¹¹ Unfortunately, it is not known whether our patient had pulmonary involvement with Kaposi's sarcoma or whether the interstitial lung disease was a direct manifestation of either his rheumatoid arthritis or polymyositis. We cannot comment on the serologic status of this patient with regard to antibodies to the human immunodeficiency virus, as the case occurred before the initial descriptions of AIDS. All of the clinical features, however, are consistent with manifestations of rheumatoid disease or polymyositis.

This case highlights the need for physicians to be aware that drug-induced immunosuppression with corticosteroids presents a significant risk factor to the development of Kaposi's sarcoma and that withdrawing the immunosuppressive therapy can result in a complete clinical remission. The skin lesions of Kaposi's sarcoma can easily be mistaken for cutaneous vasculitis, particularly in patients with autoimmune diseases on corticosteroid therapy. It is therefore important

to recognize the increased risk of Kaposi's sarcoma in such patients and to differentiate this early on from vasculitis so that therapeutic agents can be instituted or immunosuppressive therapy discontinued.

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Figure 1.—The photograph shows discrete nodular lesions over the lower extremities.

Fasciitis and Abscesses Complicating Liposuction

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SUCTION LIPECTOMY was first done in this country in 1976.¹ In the past few years the procedure has been widely publicized by the media, and the number of surgeons doing liposuction has risen dramatically.²

If patients are carefully selected, with good skin tone, localized fat deposits, and normal weight, the cosmetic results are reportedly excellent.¹⁻³ Possibly because this procedure has only recently been introduced, there has been a paucity of studies that critically examine the complication rates in large numbers of cases. The few studies available

(Bello EF, Posalski I, Pitchon H, et al: Fasciitis and abscesses complicating liposuction. *West J Med* 1988 Jun; 148:703-706)

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